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Poster session – Friday, 20 September 2013 – 12 h 00–13 h 00

16

Kommerell diverticulum should be removed when operating symptomatic children with right aortic arch and aberrant left subclavian artery (vascular ring)

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Background.— Right aortic arch with aberrant left subclavian artery is the most frequent cause of vascular ring. Usual treatment in symptomatic children is ligamentum arteriosus division, leaving the Kommerell diverticulum in place with potential risk of residual compression, aneurysmal dilation and dissection or even rupture. Translocation of the aberrant left subclavian artery to the left carotid artery together with removal of the Kommerell diverticulum and division of the ligamentum through a left thoracotomy is currently advocated to avoid those complications.

Methods and results.— Between 9/2009 and 8/2011, 13 patients underwent above-mentioned procedure. Clinical findings, surgical procedure and complications, histopathological findings and follow-up data were retrospectively analyzed. Mean age at time of surgery was 7.2 years (median 4.3, range 0.9–18.9), mean weight 25 kg (median 18, range 8.4–59). All had respiratory symptoms, associated with dysphagia in five. CT scan and/or MRI had demonstrated the arch anomaly and the dilated Kommerell diverticulum in all. A left postero-lateral thoracotomy was done in all. All had bilateral cerebral oxymetry monitoring. Postoperative complications included transient chylothorax in four and transient phrenic palsy in one patient. Mean follow-up reached 6.6 months (median 1.1, range 0.1–29). Mild residual respiratory symptoms were noted in six patients. Echo-Doppler analysis available in 11 patients showed a patent left subclavian to carotid artery anastomosis. Histopathological analysis of the resected diverticulum, available in six patients, showed cystic medial necrosis and inflammatory tissue in three, borderline cystic medial necrosis in one, hyperplastic myointimal lesions in one and nonspecific histological findings in one.

Discussion.— Translocation of the aberrant left subclavian artery together with Kommerell diverticulum resection and ligamentum division is a safe and efficient procedure for symptom relief. The observation of profound wall abnormalities such as medial necrosis in at least 50% of the analyzed diverticuli encourages us to maintain

this strategy, in order to reduce the risk of aneurysm formation and dissection.

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17

Is surgical repair of partial atrioventricular septal defect safe and efficient in adulthood?

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Background.— Partial atrioventricular septal defect (PAVSD) is usually operated in childhood with excellent long-term results. However, some patients may present during adulthood and their management as well as their outcome is still unclear. We sought to analyze the clinical characteristics at presentation and the outcome of patients with PAVSD, non-operated or operated during adulthood. **Methods.**— Between January 2000 and March 2013, 31 adult patients with PAVSD presented and were followed in our care network. The mean age at study entry was 34 (± 16.6) years.

Results.— Dyspnea was the most frequent symptom at presentation in 19 cases (61%). Left atrioventricular valve regurgitation (LAVVR) and right ventricular overload were higher in patients undergoing surgical repair ($P=0.01$). Twenty-two patients (71%) had their surgical repair at a mean age of 39.4 (± 15.3) year-old with ostium primum closure and partial or complete suture of the left atrioventricular cleft without postoperative death or major complication. There was one late reoperation for a residual shunt. Nine patients (29%) were not operated. After a mean follow-up of 7.4 (± 7.1) years, 26 patients (84%) are in NYHA class I or II whereas five (16%) are in NYHA class III or IV. Operated patients have a lower NYHA class ($P<0.01$), a lower grade of LAVVR ($P=0.03$) and a lower systolic pulmonary artery pressure ($P<0.01$) than unoperated patients at last follow-up. The onset or persistence of supraventricular arrhythmias (SVA) after surgery was associated with an older (>40 -year-old)

age at repair ($P=0.02$), a higher systolic pulmonary artery pressure ($P=0.04$) and a higher grade of LAVVR ($P=0.03$).

Conclusion.— PAVSD repair is indicated in adult patients because it carries an excellent prognosis and operated patients have better long-term clinical and hemodynamic status than unoperated patients. Moreover, SVA incidence decreases when surgery is performed before 40 years of age.

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18

Atrioventricular discordance with ventriculo-arterial concordance: Diagnostic challenge, surgical management and long-term outcome

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Background.— Atrioventricular discordance with ventriculo-arterial concordance is a rare cardiac defect whose pathophysiology resembles transposition of the great arteries.

Objective.— To report a series of ten patients with atrioventricular discordance with ventriculo-arterial concordance focusing on segmental analysis, diagnostic difficulties, surgical management and follow-up.

Methods.— Retrospective review of medical files of all patients with this diagnosis seen from 1983 to 2013 in a single institution.

Results.— Seven patients had {I, D, S} segmental arrangement, two had anatomically corrected malposition {S, L, D} and one had {S, L, S} arrangement. Only five patients were correctly diagnosed preoperatively while diagnosis had to be completed during or after surgery in the other five. Mean age at repair surgery or at last surgical intervention was 3.4 years (range: 5 months–12.8 years). Repair surgery finally performed was atrial switch procedure of Senning or Mustard type in eight of 10 patients. Repair included ventricular septal defect closure in three cases and right ventricle to pulmonary artery conduit or patch in four cases. Main postoperative complications were two cases of baffle obstruction requiring reintervention and one sick sinus syndrome needing pacemaker implantation. There was no postoperative heart block. There were two early postoperative deaths and eight late survivors. Mean follow-up after repair was 6.8 years (range: 5 months–25.4 years) with good functional status in all but one patient who will be listed for heart transplantation.

Discussion.— Preoperative diagnosis of atrioventricular discordance with ventriculo-arterial concordance remains challenging. Atrial switch procedure is the surgical method of choice. If correctly diagnosed, long-term follow-up is encouraging. Rhythm disturbances and baffle obstruction are the main postoperative problems in this series.

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19

Anomalous left coronary artery from the pulmonary artery associated with other cardiac defects: A difficult joint diagnosis

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Background.— Anomalous left coronary artery connected to the pulmonary artery (ALCAPA) is a coronary abnormality which can be associated to other congenital heart defects which complicates the positive diagnosis of the coronary abnormality, especially before surgery.

Objective.— Here, we report a series of 13 patients with ALCAPA with a focus on the type of associated heart defect, the moment of diagnosis of the coronary abnormality related to surgery and their outcome.

Methods.— Retrospective assessment of medical files of all patients with ALCAPA and other congenital heart defects in two important French Departments of Pediatric Cardiac Surgery from 1987 to 2012.

Results.— The heart defect most frequently encountered in association to ALCAPA was aortic coarctation ($n=5$) followed by tetralogy of Fallot with or without pulmonary atresia ($n=3$). There was one case of hypoplastic left heart syndrome, one aortic atresia, one right aortic arch, one congenital mitral malformation and one infant with divided left atrium and partial anomalous pulmonary venous return of the right lung. Only four patients had a complete diagnosis of the cardiac defect and the coronary abnormality before surgery. In three cases the coronary anomaly was discovered during surgery conducted for another cardiac defect and treated at the same time by coronary reimplantation. The six remaining patients were diagnosed after cardiac repair. Three of these patients only had a post-mortem diagnosis. Eight of 13 patients died after surgery. Three of them deceased within the first 30 days after repair. The remaining patients are in good health with a median follow-up of 5.3 years (range: 2.1–8.5 years).

Discussion.— This series confirms that ALCAPA associated with other cardiac defects is often unrecognized before surgery. Pulmonary hypertension due to left-to-right shunt or coarctation can maintain an antegrade flow in the anomalous coronary artery until cardiac repair. Myocardial ischemia will only become apparent once the defect has been repaired when pulmonary pressure lowers. In this series postoperative survival was compromised mostly due to complications occurring after complex or repeated cardiac surgery.

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20

The medical past of adults with complex congenital heart disease impacts their social development and professional activity

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Advances in surgery and therapeutic catheterization in recent decades increased steadily life expectancy and prevalence of adult patients with congenital heart disease (ACHD). We assessed medical and psychosocial parameters of ACHD according to the complexity of the disease.

Methods.— We included from a single-center prospective observational cohort study which started in January 2013, 68 ACHD patients (40.7±13.9 years old, max 77) followed in cardiology unit from January to April 2013 who answer a questionnaire assessing daily activity and psychosocial being. Complexity of the disease was classified according to the 32nd Bethesda conference.

Results.— Cardiac malformations were simple (S) in 30 patients (44.1%), moderate (M) in 26 (38.2%) and complex (C) in 12 (17.7%). Pediatric cardiac surgery had been performed in 84.2% of M and C